PURE KYPHOSIS IN NEUROFIBROMATOSIS. A CASE REPORT

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Introduction

Deformity of the spine is common in patients with neurofibromatosis, especially scoliosis and kyphoscoliosis. An almost pure progressive thoracic kyphotic deformity has rarely been reported.

When severe kyphosis is associated with mild scoliosis, attention must be directed to the kyphotic element of deformity as it alone usually contributes to paraplegia[3,11]. Paraplegia in neurofibromatosis may also result from vertebral subluxation or dislocation, or from intraspinal tumor.

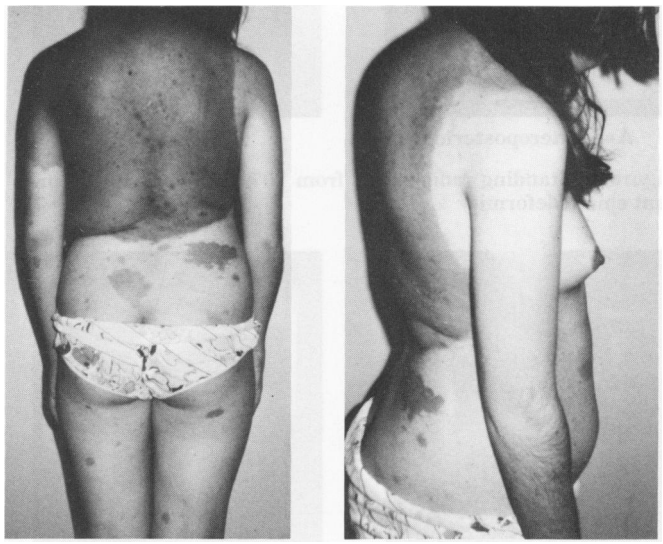
Case

N.B., a nineteen year, three month old white female with histologically proven neurofibromatosis, presented with increasing back pain. Her father and four of five siblings had the same disease. She had a several year history of increasing, constant, burning type interscapular pain, exacerbated by activity and decreased slightly with rest. She complained of occasional headaches although denied radicular pain in her arms or legs.

Past history included a right both bone forearm fracture which healed after three to four years and multiple operations and bone graftings.

Examination revealed multiple cafe-au-lait spots scattered over her body. She stood with a receding chin and forward tilt to her head along with an increased thoracic kyphosis and lumbar lordosis (Figs. 1A and 1B). The left shoulder was elevated one centimeter compared to the right shoulder. A plumbline dropped from the seventh cervical spinous process fell one centimeter to the right of the mid-gluteal crease. The pelvis was level. On forward bending, there was a left thoracic rib prominence of 0.4 centimeters as well as an increased thoracic kyphosis (Fig. 2). Neurological examination was normal.

Old standing roentgenograms are shown in Figures 3A and 3B. New roentgenograms (Figs. 4A and 4B) revealed a 12 degree left thoracic curve from T5 to T12 and a 10 degree right lumbar curve from T12 to L3. The lateral roentgenogram revealed a thoracic kyphosis from T2 to T8 of 103 degrees with the apex at T5-6. A hyperextension view showed little correction of the kyphosis.



A—Back B—Side

Figure 1. Preoperative photographs

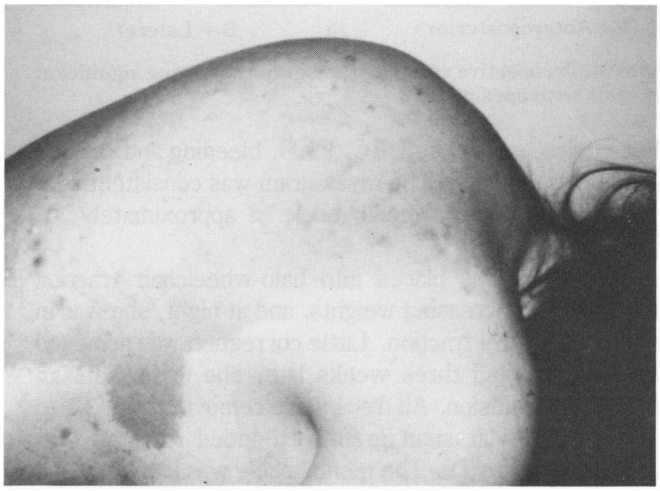


Figure 2. Lateral forward bending preoperative photograph

Course in the Hospital

Pulmonary functions revealed an FEV of 64.5 per cent, an FEV_1 of 82.5 per cent and a PaO_2 of 94.5 per cent.

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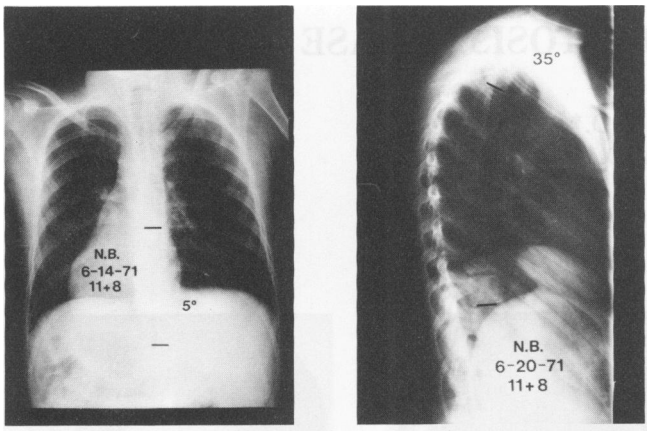
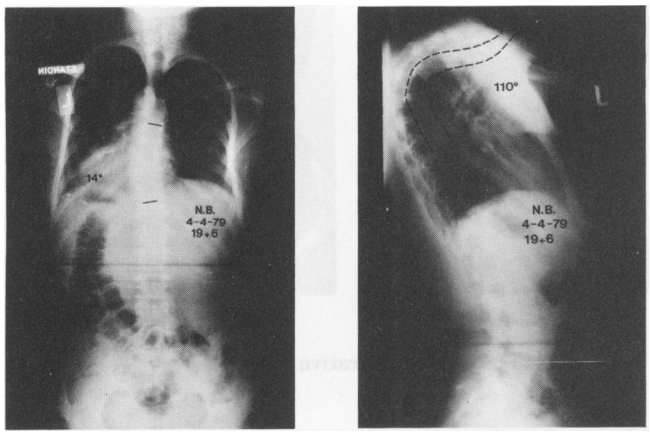


A—Anteroposterior B—Lateral

Figure 3. Standing radiographs from 1971 showing no significant spinal deformity



A—Anteroposterior B—Lateral

Figure 4. Preopertive standing radiographs showing significant kyphosis with apex at T5-6

Chemistries, enzymes, CBC, EKG, bleeding and clotting studies were normal. The myelogram was consistent with a left lateral thoracic meningocele at approximately T5 (Fig. 5).

The patient was placed into halo-wheelchair traction with gradually increasing weights, and at night, she was in bed in halo-Cotrel traction. Little correction was achieved with traction, and three weeks later she underwent an anterior spinal fusion. All discs were removed from T1 to T10, and a fibular strut graft with added rib graft was placed from T1 to T9. The meningocele was identified and care was taken to avoid puncturing the dural sac. She was maintained in halo traction postoperatively. Approximately four weeks later she underwent a posterior spinal fusion employing two Harrington compression rods. Since the upper four 1259 hooks were placed over weak transverse processes, these hooks were secured to the spine with wire and methylmethacrylate. Ten days postoperatively, she was placed into a halo-Risser cast, ambulated and

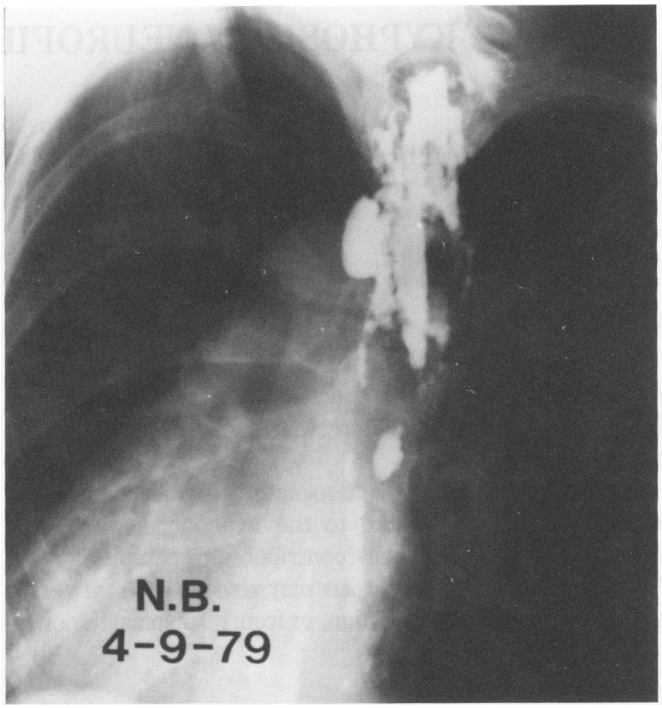
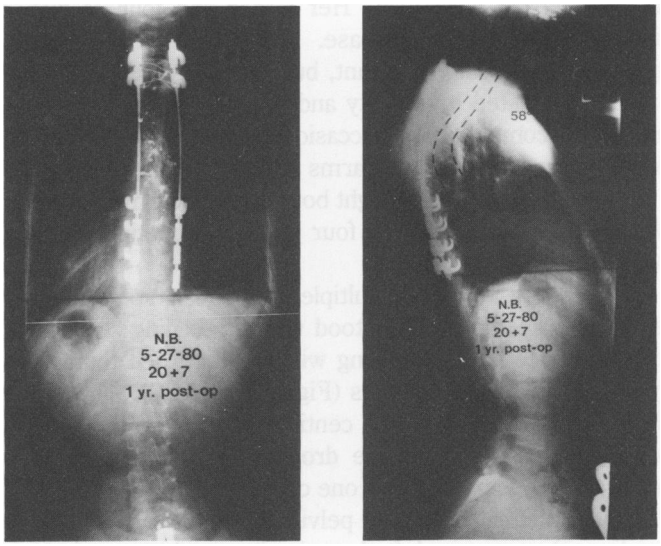


Figure 5. Preoperative myelogram showing lateral thoracic meningocele

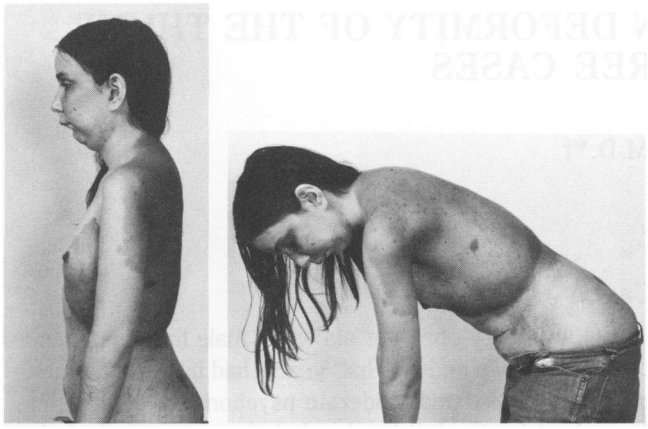


A—Anteroposterior B—Lateral

Figure 6: One year postoperative radiographs

discharged. She remained in the halo cast for five months and was then placed in a full Risser-Cotrel cast for six months. Postoperative radiographs are seen in Figures 6A and 6B. The clinical photographs are seen in Figures 7A and 7B.

At two years and nine months following fusion, full correction has been maintained. No pseudarthroses have been identified. Complete relief of back pain was achieved.



A — Side view B — Forward bending

Figure 7. Postoperative photographs

Discussion

Some of the previous reports on the occurrence of kyphosis in neurofibromatosis are lacking in detail. Rezaian mentioned two cases of kyphosis but with minimal discussion[8]. Hagelstam reported twelve cases of kyphosis, but the majority were a gibbus, probably due to tuberculosis[4]. Henyer and Feld reported a case of thoracic kyphosis with mild scoliosis associated with paraplegia[5]. However, it is unclear if the deformity was present prior to laminectomy or arose postoperatively.

One case similar to ours was reported by Kessel in 1951[7]. His patient, twenty-three years of age, had increased thoracic kyphosis with little scoliosis and had a lateral intrathoracic meningocele noted at surgery. No further cases were found in the English literature.

The presence of a lateral intrathoracic meningocele was first reported in 1933[10]. By 1972, thirty-seven cases had been reported, twenty-eight of which were associated with neurofibromatosis[1]. The diagnosis of lateral intrathoracic meningocele is difficult since there are usually no neurological symptoms. Meningoceles have been reported at the apex of a scoliosis on the convex side[9], and since many curves are right sided, most meningoceles occur in the right hemithorax[1,6]. The etiology of the meningocele is unknown, although it has been proposed to be secondary to a posterior neural arch defect[10]. Specific treatment of the meningocele does not seem to be indicated unless neurologic findings are present. There have been no sequelae from the meningocele in this case.

As seen in this case, severe kyphosis can develop despite the lack of significant progressive scoliosis. If the spine is viewed only in two dimensions, progressive kyphotic or lordotic curves will be overlooked.

Summary

A case report of severe thoracic kyphosis without significant scoliosis in neurofibromatosis was presented. The treatment and associated findings were discussed.

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